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January 27, 2007

State and Provincial Updates

Alberta:
Alberta is continuing testing for the provincial chronic wasting disease (CWD) surveillance program for 2006-07. One additional case of CWD in wild deer was confirmed on January 2. This brings the total to 17 cases in wild deer in Alberta since the first case in September 2005. The most recent case was taken by a hunter near Chauvin, not far from an earlier case reported in late December.
Alberta Sustainable Resource Development's CWD information is at:
http://www.srd.gov.ab.ca/fw/diseases/CWD/index.html.
A map showing the locations of Alberta CWD+ animals is at:
http://www.srd.gov.ab.ca/fw/diseases/CWD/pdf/CWD_positive_Dec2006.pdf.

West Virginia:
On December 22, 2006 the West Virginia DNR announced that a hunter-harvested deer taken during the 2006 hunting season tested positive for CWD. The 2.5 year old buck, one of 1355 deer sampled in Hampshire County during the 2006 hunting season, was taken in close proximity to the previously-established cluster of CWD+ deer in Hampshire County. CWD has now been detected in a total of 10 deer in West Virginia, all in Hampshire County: one road-killed deer, four deer collected by the DNR in 2005, four deer collected by the DNR in 2006 and one hunter-harvested deer during the 2006 deer season.
West Virginia DNR CWD information is at: http://www.wvdnr.gov/hunting/ChronicWaste.shtm.
A WVDNR fact sheet is at: http://www.wvdnr.gov/hunting/PDFFiles/CWDfactsheet.pdf.

Wisconsin:
The Wisconsin DNR has begun agency sharpshooting at Devil's Lake State Park and the Southern Unit of Kettle Moraine State Forest. Devil's Lake S.P. is the furthest north that CWD has been detected in the state; three CWD+ deer have been detected in the 10,200 acre park in Sauk County in the last year. Kettle Moraine State Forest lies just north of the southeastern CWD zone in Wisconsin; five hunter-harvested deer from the 6400 acre Walworth County portion of the forest tested positive this year. In an additional note, the DNR has begun conducting this year's winter helicopter deer counts in CWD-affected areas. Recent snowfall and cold temperatures are providing good conditions for these efforts to determine deer density within Wisconsin's CWD zones.
Wisconsin Department of Natural Resources CWD information, including an interactive map showing locations of positives and overall surveillance effort, is at:
http://www.dnr.state.wi.us/org/land/wildlife/whealth/issues/CWD/index.htm.

Wyoming:
The Wyoming Game and Fish Department has finished its fourth year of comprehensive Chronic Wasting Disease surveillance and added two deer hunt areas and two elk hunt areas to its list of areas where CWD has been detected. Game and Fish personnel collected 4,653 deer, elk and moose samples in 2006. Of those, 116 animals tested positive for CWD – 88 mule deer, 13

white-tailed deer and 15 elk. New cases of CWD were diagnosed in deer hunt area 4 east of Sundance, deer hunt area 11 in Niobrara and Weston counties and elk hunt areas 16 and 22 in northern Carbon County.
The Wyoming Game & Fish Department 2006 surveillance summary press release is at: http://gf.state.wy.us/services/news/pressreleases/06/12/15/061215_2.asp.
Wyoming Game & Fish Department CWD Information is at: http://gf.state.wy.us/services/education/cwd/index.asp.

Recent Publications

Production of cattle lacking prion protein.
Jürgen A Richt, Poothappillai Kasinathan, Amir N Hamir, Joaquin Castilla, Thillai Sathiyaseelan, Francisco Vargas, Janaki Sathiyaseelan, Hua Wu, Hiroaki Matsushita, Julie Koster, Shinichiro Kato, Isao Ishida, Claudio Soto, James M Robl & Yoshimi Kuroiwa
Nature Biotechnology 25, 132-138 (2006).

Abstract: Prion diseases are caused by propagation of misfolded forms of the normal cellular prion protein PrP^{C}, such as PrP^{BSE} in bovine spongiform encephalopathy (BSE) in cattle and PrP^{CJD} in Creutzfeldt-Jakob disease (CJD) in humans. Disruption of PrP^{C} expression in mice, a species that does not naturally contract prion diseases, results in no apparent developmental abnormalities. However, the impact of ablating PrP^{C} function in natural host species of prion diseases is unknown. Here we report the generation and characterization of PrP^{C}-deficient cattle produced by a sequential gene-targeting system. At over 20 months of age, the cattle are clinically, physiologically, histopathologically, immunologically and reproductively normal. Brain tissue homogenates are resistant to prion propagation in vitro as assessed by protein misfolding cyclic amplification. PrP^{C}-deficient cattle may be a useful model for prion research and could provide industrial bovine products free of prion proteins.
http://www.nature.com/nbt/journal/v25/n1/abs/nbt1271.html

The role of the cellular prion protein in the immune system.
J. D. Isaacs, G. S. Jackson, and D. M. Altmann
Clinical & Experimental Immunology 146 (1), 1–8 (2006).

Abstract: Prion protein (PrP) plays a key role in the pathogenesis of prion diseases. However, the normal function of the protein remains unclear. The cellular isoform (PrP^{C}) is expressed widely in the immune system, in haematopoietic stem cells and mature lymphoid and myeloid compartments in addition to cells of the central nervous system. It is up-regulated in T cell activation and may be expressed at higher levels by specialized classes of lymphocyte. Furthermore, antibody cross-linking of surface PrP modulates T cell activation and leads to rearrangements of lipid raft constituents and increased phosphorylation of signalling proteins. These findings appear to indicate an important but, as yet, ill-defined role in T cell function. Although $PrP^{-/-}$ mice have been reported to have only minor alterations in immune function, recent work has suggested that PrP is required for self-renewal of haematopoietic stem cells. Here, we consider the evidence for a distinctive role for PrP^{C} in the immune system and what the effects of anti-prion therapeutics may be on immune function.
http://www.blackwell-synergy.com/doi/abs/10.1111/j.1365-2249.2006.03194.x.

Prions and their partners in crime.
Byron Caughey and Gerald S. Baron
Nature 443, 803-810 (19 October 2006)

Abstract: Prions, the infectious agents of transmissible spongiform encephalopathies (TSEs), have defied full characterization for decades. The dogma has been that prions lack nucleic acids and are composed of a pathological, self-inducing form of the host's prion protein (PrP). Recent progress in propagating TSE infectivity in cell-free systems has effectively ruled out the involvement of foreign nucleic acids. However, host-derived nucleic acids or other non-PrP molecules seem to be crucial. Interactions between TSE-associated PrP and its normal counterpart are also pathalogically important, so the physiological functions of normal PrP and how they might be corrupted by TSE infections have been the subject of recent research.
http://www.nature.com/nature/journal/v443/n7113/abs/nature05294.html.

The following review article, authored by Beth Williams, was published in 2005, following her unfortunate and untimely death. While aspects of our knowledge regarding CWD have been updated by more recent science, this review article is probably the most thorough ever written and is highly recommended to bolster the reader's overall understanding of CWD.

Chronic Wasting Disease.
E. S. Williams
Veterinary Pathology 42:530–549 (2005)

Abstract: Chronic wasting disease (CWD) is a unique transmissible spongiform encephalopathy (TSE) of mule deer (Odocoileus hemionus), white-tailed deer (O. virginianus), and Rocky Mountain elk (Cervus elaphus nelsoni). The natural history of CWD is incompletely understood, but it differs from scrapie and bovine spongiform encephalopathy (BSE) by virtue of its occurrence in nondomestic and free-ranging species. CWD has many features in common with scrapie, including early widespread distribution of disease-associated prion protein (PrP^d) in lymphoid tissues, with later involvement of central nervous system (CNS) and peripheral tissues. This distribution likely contributes to apparent efficiency of horizontal transmission and, in this, is similar to scrapie and differs from BSE. Clinical features and lesions of CWD are qualitatively similar to the other animal TSEs. Microscopically, marked spongiform lesions occur in the central nervous system (CNS) after a prolonged incubation period and variable course of clinical disease. During incubation, PrP^d can be identified in tissues by antibody-based detection systems. Although CWD can be transmitted by intracerebral inoculation to cattle, sheep, and goats, ongoing studies have not demonstrated that domestic livestock are susceptible via oral exposure, the presumed natural route of exposure to TSEs. Surveillance efforts for CWD in captive and free-ranging cervids will continue in concert with similar activities for scrapie and BSE. Eradication of CWD in farmed cervids is the goal of state, federal, and industry programs, but eradication of CWD from free-ranging populations of cervids is unlikely with currently available management techniques.
http://www.vetpathology.org/cgi/content/abstract/42/5/530.

Upcoming Conferences

Transmissible Spongiform Encephalopathies: The Definitive American TSE Meeting
February 12-13, 2007, Baltimore, Maryland
http://www.healthtech.com/2007/tse/index.asp